

1/5

<graphic>

Key:

BH<sub>4</sub>-sensitiv = BH<sub>4</sub> sensitive

Milde = mild

Plasma-phenylalanin = Plasma Phenylalanine

Vor = Before

H = hours

<graphic>

Key:

BH<sub>4</sub>- nicht sensitiv = BH<sub>4</sub> nonsensitive

Milde = mild

Plasma-phenylalanin = Plasma Phenylalanine

Vor = Before

H = hours

Drawing 1

2.1/5

A

<graphic>

**Key:**

BH<sub>4</sub>-sensitiv = BH<sub>4</sub> sensitive

Milde = mild

Kumulative <sup>13</sup>CO<sub>2</sub> recovery = Cumulative <sup>13</sup>CO<sub>2</sub> recovery

% der Dosis = % of Dose

Vor = Before

Nach = After

<graphic>

**Key:**

BH<sub>4</sub>-nicht sensitive = BH<sub>4</sub> insensitive

Milde = mild

Kumulative <sup>13</sup>CO<sub>2</sub> recovery = Cumulative <sup>13</sup>CO<sub>2</sub> recovery

% der Dosis = % of Dose

Vor = Before

Nach = After

**Drawing 2**

2.2/5

<graphic>

**Key:**

BH<sub>4</sub>-sensitive = BH<sub>4</sub> sensitive

Milde = mild

Zeit (min) = Time (min)

BH<sub>4</sub>-nicht sensitive = BH<sub>4</sub> insensitive

**Drawing 2, Continued**

3/5

<graphic>

**Key:**

Vor = Before

Nach = After

Plasma-phenylalanin = Plasma Phenylalanine

Kumulative  $^{13}\text{CO}_2$  recovery = Cumulative  $^{13}\text{CO}_2$  recovery

**Drawing 3**

4/5

<graphic>

**Key:**

Plasma-phenylalanin = Plasma Phenylalanine

Kumulative  $^{13}\text{CO}_2$  recovery = Cumulative  $^{13}\text{CO}_2$  recovery

BH<sub>4</sub>-sensitive = BH<sub>4</sub> sensitive

Moderat BH<sub>4</sub>-sensitive = Moderately BH<sub>4</sub> sensitive

BH<sub>4</sub>-nicht sensitive = BH<sub>4</sub> insensitive

**Drawing 4**

5/5

<graphic>

**Key:**

Regulatorische Domäne = Regulatory Domain

katalytische Domäne = Catalytic Domain

Tetramerisierungsdomäne = Tetramerization Domai

**Drawing 5**